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Review Article

Double-chambered right ventricle: a review

Marios Loukas [a,*], Brian Housman [a], Christa Blaak [a], Sarah Kralovic [a], R. Shane Tubbs [b], Robert H. Anderson [c]

[a] Department of Anatomical Sciences, School of Medicine, St George's University, Grenada, West Indies
[b] Children's Hospital, Pediatric Neurosurgery, Birmingham, AL, USA
[c] Institute of Genetic Medicine, Newcastle University, Newcastle, United Kingdom

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ABSTRACT

A double-chambered right ventricle is a rare heart defect in which the right ventricle is separated into a high-pressure proximal and low-pressure distal chamber. This defect is considered to be congenital and typically presents in infancy or childhood but has been reported to present rarely in adults. It can be caused by the presence of anomalous muscle tissue, hypertrophy of the endogenous trabecular bands, or an aberrant moderator band; all of which will typically result in progressive obstruction of the outflow tract. In this paper, we will discuss the general anatomy of the right ventricle, the relevant embryology of the heart, and the presentation, diagnosis, and treatment of a double-chambered right ventricle.

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1. Introduction

A double-chambered right ventricle (DCRV) is a heart defect, typically congenital, in which the right ventricle (RV) is separated into a proximal high-pressure (anatomically lower) chamber and distal low-pressure (anatomically higher) chamber [1]. It can be caused either by the presence of anomalous muscle bands, by hypertrophy of endogenous trabecular tissue, or occasionally by an aberrant moderator band. DCRV is frequently associated with other congenital heart defects, chiefly a membranous ventricular septal defect, but also tetralogy of Fallot, Ebstein's anomaly, and double outlet RV, among many others [2]. In cases such as anomalous musculature or an atypical moderator band, DCRV may be considered a cardiac congenital defect as well. However, DCRV has also been reported to develop postnatally due to progressive hypertrophic changes in the crista supraventricularis or other muscular structures within the RV [3,4].

DCRV was first described by Peacock in 1867 as a constriction of the proximal portion of the infundibulum [5]. In 1909, Keith described a muscular shelf extending into the apex of the ventricle. Brock later described, in 1957, an infundibular muscular obstruction in the setting of tetralogy of Fallot [6,7]. Outflow obstruction was observed to be directly caused by anomalous muscle tissue by Tsifutis in 1961 and was first surgically corrected by Lucas et al. in 1962 through a partial ventriculotomy [8,9].

DCRV is characterized by intraventricular pressure gradients greater than 20 mmHg, turbulent flow patterns in the ventricle, and increased pulmonary flow [2]. However, the methods for detecting these changes vary significantly in both efficacy and accuracy [10,11]. An understanding of the anatomy and embryological development of the heart is a key in understanding the pathology of DCRV, as well as its diagnosis and treatment.

2. General anatomy of the RV

There is still substantial disagreement regarding terminology of the morphological structures of the RV. In order to simplify our review of double-chambered pathology, we will rely on the descriptions and terms outlined by Anderson et al. [12] and Wilcox et al. [13].

The RV is composed of three anatomically distinct regions: the inlet, the trabecular apex, and the outlet [13]. The inlet, or inflow tract, includes, and is limited by, the tricuspid valve. True to its name, the tricuspid is composed of three leaflets, the anterosuperior, inferior, and septal. These are tethered peripherally by fan-shaped chordae tendinea, which are then anchored by the papillary muscles into the

* Corresponding author. Department of Anatomical Sciences, St. George's University, School of Medicine, Grenada, West Indies. Tel.: +1 473 444 4175x2005; fax: +1 473 444 2887.
E-mail address: mloukas@sgu.edu (M. Loukas).

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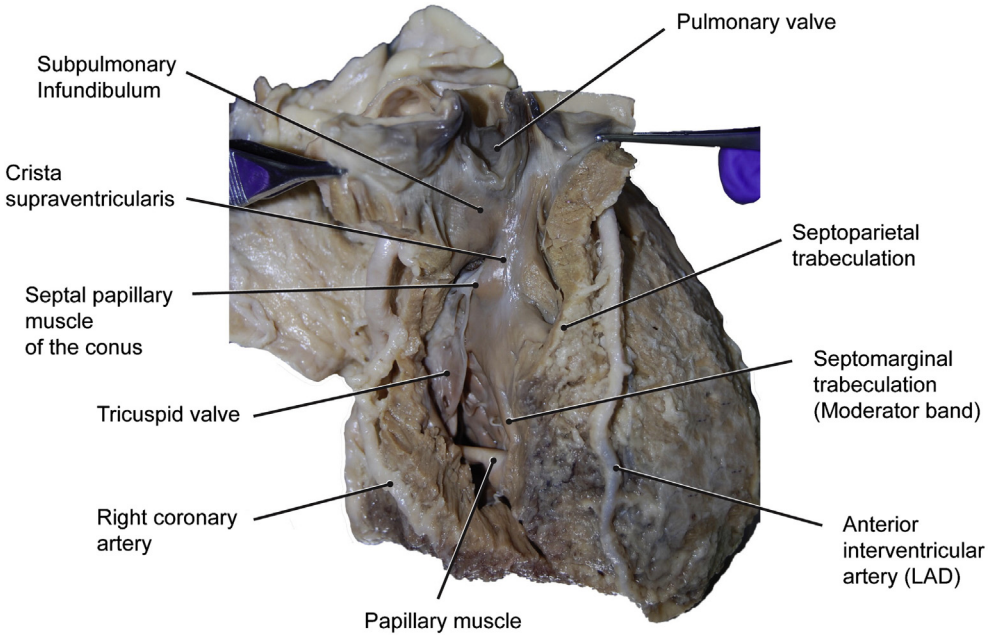

Fig. 1. This figure is taken from a normal dissected RV exhibiting key anatomical landmarks.

trabeculated myocardial wall, except for the cords of the septal leaflet, which are directly attached to the interventricular septum via the septal papillary muscle of Luschka [13,14]. The trabecular portion of the ventricle extends to the apex where the wall is particularly thin and prone to injury by cardiac catheters and pacing electrodes [13].

The outlet, or outflow tract, is characterized by smooth, muscular walls. It is the only nontrabeculated region of the RV, as both the inlet and apex contain numerous, coarse trabeculations [13,15]. The outlet is composed of the smooth, muscular infundibulum, which divides the aorticopulmonary trunk. It extends from the base of the RV as a muscular sleeve that supports the leaflets of the pulmonary valve and surrounds and supports the base of the pulmonary trunk [13]. Since the aorticopulmonary septum may be displaced in anomalous hearts, Anderson et al. [12] proposed that the term infundibular septum be used to describe any structure that separates two semilunar valves.

Two other morphologically distinct characteristics of the RV include the supraventricular crest, which is clasped by a prominent trabecula known as the septomarginal trabeculation. The supraventricular crest, or crista supraventricularis, is a muscular shelf that normally separates the tricuspid and pulmonary valves [13,15]. This structure often appears to be a bundle of muscle but is actually an infolding of the inner curve of the heart, known as the ventriculo-infundibular fold. This fold inserts into a prominent and important septal trabeculation, located toward the apex of the ventricle, known as the septomarginal trabeculation [13]. Anderson et al. [12] suggested that the term crista supraventricularis should only be used in the context of normal right ventricular outflow tracts (RVOTs), when no anomalies misalign the outflow musculature. In the context of anomalous hearts, the term ventriculo-infundibular fold can be used to identify any structure that separates an atrioventricular valve from a semilunar valve [12].

The septomarginal trabeculation is a prominent Y-shaped band that is continuous with the interventricular septum inferiorly and towards the base of the heart, gives rise to an anterosuperior limb and a posteroinferior limb (Fig. 1). The anterosuperior, or septal, limb migrates superiorly to overlie the outlet part of the muscular ventricular septum. Here, it supports the pulmonary valve in the outlet of the ventricle. The posteroinferior, or parietal, limb extends posteroinferiorly to the interventricular component of the membranous septum, reinforcing this muscular structure [13]. The body of the septomarginal trabeculation continues down into the apex of the RV, breaking into several smaller, but prominent, trabeculations; most of these are termed simply “septoparietal bands” [3,13]. One of these trabeculations is particularly prominent: the moderator band. This band attaches to both the apex and base of the interventricular septum, then joins to support the anterior papillary muscle before crossing over to the free wall of the ventricle [13,16]. The moderator band is a useful landmark in echocardiography, as it allows the operator to identify the RV, particularly when it is hypertrophied or dilated [17]. Even in normal hearts, the moderator band is found in variable positions throughout the RV [5,16]. The various septoparietal bands are also quite variable, especially in size, and some may appear large enough to be mistaken for the moderator band [5].

3. Etiology

DCRV is a rare condition seen in only 0.5–2.0% of all cases of congenital heart disease and is most frequently encountered in infants and children [2,3,18]. While cases have been found in adults, these might be due to missed diagnoses during infancy rather than novel onset later in life [2,11]. Alternatively, since cases of DCRV are frequently associated with various other congenital cardiac defects, it is also possible that the pathology develops as a result of turbulent flow in the ventricular outlet, causing hypertrophy of muscular tissue within the RV [2].

Since DCRV is considered a primarily congenital pathology, a short discussion on the embryology of cardiac development is necessary. The majority of cardiac structures derive from a primary heart tube, situated centrally within the embryo as an inverted Y. The inferior limbs of the heart tube go on to form the atrial chambers, while the stem forms the left ventricle (LV) [19]. The RV and outflow tract develop from a secondary heart field, located medial to, but contiguous with, the primary heart tube. The primary heart tube undergoes looping ventrally and to the right, creating an inner and an outer curvature, which gives rise to inlet and outlet components, respectively [19]. The apical parts of the two ventricles balloon out from the outer curvature of this loop, the inlet part giving rise to the LV and the outlet part giving rise to the RV [19] (Fig. 2).

Endocardial cushion cells form on the superior and inferior aspects of the atrioventricular canal, which fuse together to form the right and

left components of the canal [21]. Cushions are complex tissues that contain epithelium, mesenchyme, and extracellular material [22]. The muscular ventricular septum develops concomitantly with the ballooning of the ventricles, and as the divided atrioventricular canal expands, the inferior atrioventricular cushion becomes "draped" over the right side of the posteroinferior portion of the muscular septum [21]. The fused endocardial cushion cells go on to form the central fibrous body of the cardiac skeleton [21]. Cushion cells also develop more proximally, in the outflow tract of the developing heart [22]. Fused outflow cushion cells become muscularized and incorporated into the RV to become the subpulmonary infundibulum and the supraventricular crest [21].

Due to the interconnected nature and complexity of these developing structures, it stands to reason that a congenital defect resulting in malformation of the outflow tract could substantially affect the development of the interventricular septum and crista supraventricularis. It is therefore understandable that there are multiple embryological and postnatal causes of DCRV.

4. Clinical definition of DCRV

DCRV is typically considered to be a congenital heart disorder caused by anomalous muscle dividing the RV [23] (Fig. 3). Restivo et al. [3] acknowledged that the terms "anomalous right ventricular muscle bundle" and "two-chambered right ventricle" are frequently used synonymously, and countered this assumption by presenting their observations that a variety of lesions can cause division of the RV. The reports of Restivo et al. [3] defined the seven most commonly cited morphologies of the divided RV. Each of these cases falls into two main categories of obstruction: anomalous muscle tissue and hypertrophic endogenous trabeculae. Park et al. [11] classified these groups as Type 1 DCRV, characterized by anomalous muscle tissue crossing the RV, and Type 2 DCRV, characterized by parietal and septal muscle hypertrophy.

Alva et al. [5] described the anomalous muscle tissue as septoparietal trabeculations, which have been displaced within the RV. These are contrasted with myocardial bands that have normal attachments but have undergone marked hypertrophic changes [5]. Wong et al., in 1991, suggested that DCRV may also result from a moderator band with a more superior origin, or "high take-off," and almost complete detachment from the septal wall [5,16]. Reports of DCRV resulting from neoplastic obstruction have not been identified. No genetic pattern for DCRV has been identified [24].

Many authors have reported a high incidence of concomitant congenital disease; up to 90% of cases are associated specifically with membranous-type ventricular septal defects [2]. Other reported coexistent conditions include pulmonary valve stenosis, anomalous pulmonary drainage, transposition of the great arteries, pulmonary atresia, atrial septal defect, aortic valve regurgitation, tricuspid valve regurgitation, persistent left superior vena cava, double outlet RV, ruptured sinus of Valsalva aneurysm, tetralogy of Fallot, and Ebstein's anomaly [2,11]. Ebstein's anomaly can itself result in division of the RV if it is sufficiently large and angled inferiorly across the inlet [3]. The tendency of DCRV to present in conjunction with other congenital cardiac defects, and rarely arise in isolation, suggests a strong correlation with a developmental origin [2].

However, cases of de novo DCRV have been reported. Folger [18] described multiple patients with new-onset intraventricular pressure gradients, along with the emergence of a contractile pouch in the outflow tract. These cases suggested that postnatal flow patterns within the RV might cause hypertrophic changes in the trabeculae. Maron et al. [25] reported that increased flow or pressure may stimulate hypertrophic changes of the crista supraventricularis. Massin [26] described the potential development of iatrogenic DCRV following the repair of a ventricular septal defect. Prior to surgery, no pressure gradient was detected, and no muscle proliferation was seen on echocardiography. However, postoperative follow-up revealed hypertrophy of the moderator band and a pressure gradient across the RV, which reached up to 60 mmHg [26]. These findings illustrate the importance of considering DCRV in patients with ventricular septal defect, even following successful surgical or primary closure of the septal defect.

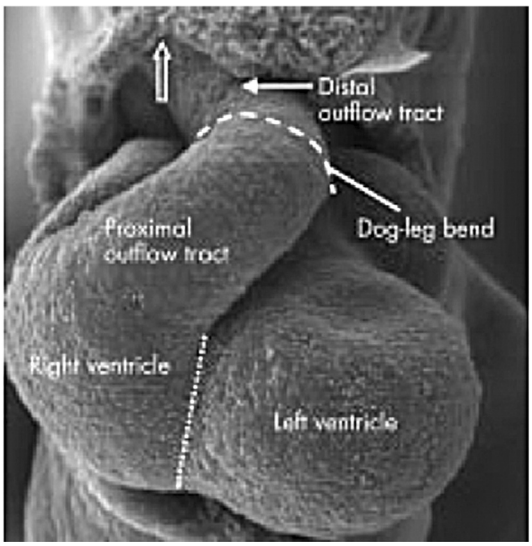

Fig. 2. This scanning electron micrograph shows the heart from a mouse with 42 somites. The front of the pericardial cavity has been removed. The outflow tract is supported exclusively by the RV, the dotted line showing the location of the interventricular groove. Note the characteristic bend (dashed line) dividing the outflow tract into proximal and distal portions. Note also that the distal portion extends to the margins of the pericardial cavity (arrow). Reproduced from Anderson et al. [20] with permission from *Heart*. Copyright 2003.

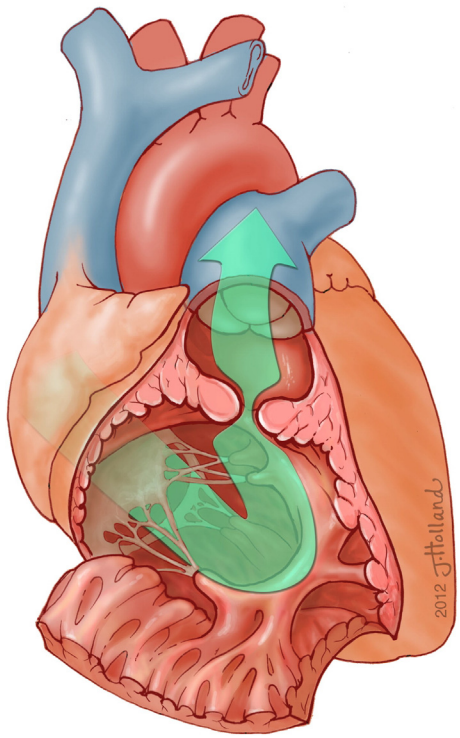

Fig. 3. This illustration exhibits the abnormal muscular tissue contributing to the cause of DCRV.

Many diseases can cause intraventricular pressure changes, it is therefore important to distinguish DCRV from other forms of outflow tract obstruction. DCRV is the only RVOT obstruction classified as subinfundibular. Most other causes of outflow obstruction occur either at or above the valve, at the infundibulum, or iatrogenically [24]. However, since pathologic turbulent flow may act as the initial stimulus for hypertrophic changes to the trabeculae, acquired DCRV may be the inevitable consequence of any outflow obstruction, causing progressive hypertrophy of the crista supraventricularis.

The distinction of etiology is further clouded by the observation that DCRV is frequently misdiagnosed and may persist asymptomatically into adulthood [2,10]. The characteristics of the pathology suggest that DCRV may be more aptly classified as an extreme symptom of other cardiac diseases rather than as an independent pathology. Aschcraft et al. [27] stated that anomalous muscle bands in the RV may actually be quite common and that it is only in the presence of congenital heart defects causing right ventricular hypertrophy that these anatomical anomalies cause symptomatic outflow obstruction.

5. Diagnosis of DCRV

DCRV shares diagnostic features with many other forms of heart disease. Patients have been reported to present with dyspnea, syncope, both stable and unstable angina, and exercise intolerance [10,11,28,29]. Left-sided parasternal systolic murmurs have been auscultated in DCRV, which are typically displaced lower along the sternal border than a typical systolic murmur [29,30]. However, findings of a murmur are far from conclusive [2,11]. In cases of marked pulmonary outlet obstruction, right ventricular hypertrophy may result. Such cases may present with a right ventricular heave, which may be accompanied by a palpable thrill [24].

Electrocardiogram testing on patients may reveal right ventricular hypertrophy. Right axis deviation, prominent R waves in the right precordial leads, and an absence of prominent S waves in the left precordial leads have all been reported in cases of DCRV and suggest right ventricular hypertrophy [10,31]. Signs of right ventricular hypertrophy are an important diagnostic clue, but a more detailed investigation is necessary since many pathologies can cause hypertrophy. A diagnosis of DCRV should be considered when right ventricular hypertrophy is apparent with no signs of infundibular hypertrophy or valvular pulmonary stenosis [23].

The most effective form of diagnosis of DCRV is transesophageal echocardiography [2] (Fig.4A). In the context of echocardiography, obstructions are defined in several ways based on their location and appearance. A lesion is described as “high” when situated adjacent to the pulmonary valve and “low” if localized in the apex. “Diffuse” lesions are triangular with long attachments and “discrete” lesions are well defined with short attachments [2]. The transesophageal approach with Doppler allows for visualization of the degree of stenosis between the two chambers [5] (Fig.4B,C). Use of continuous Doppler also allows for the calculation of intraventricular pressure gradients and obviates the need for additional cardiac catheterization [2]. On echocardiography, fibrotic and hypertrophic tissue is more evident during systole, since the muscle bands are contracted and thickened [23].

Compared to the transesophageal approach, transthoracic echocardiography is less accurate. Hoffman et al. [2] reported that in a study of 32 adult patients with DCRV, the transthoracic approach was only diagnostic in 5 out of 32 studies; whereas, transesophageal echocardiography accurately diagnosed DCRV in all 21 cases for which it was used. Transthoracic echocardiography is typically used successfully in pediatric patients [32]. However, in adults, a transesophageal approach provides better definition of congenital lesions [2]. Romano et al. [32] suggested that limitations in transthoracic echocardiography are primarily due to the proximity of the RVOT to the transducer on the precordium. Other limitations, particularly encountered in adults, include obesity and emphysema [32]. While warning of its limited diagnostic potential, Hoffman et al. [2] recommended that when using a transthoracic approach, the

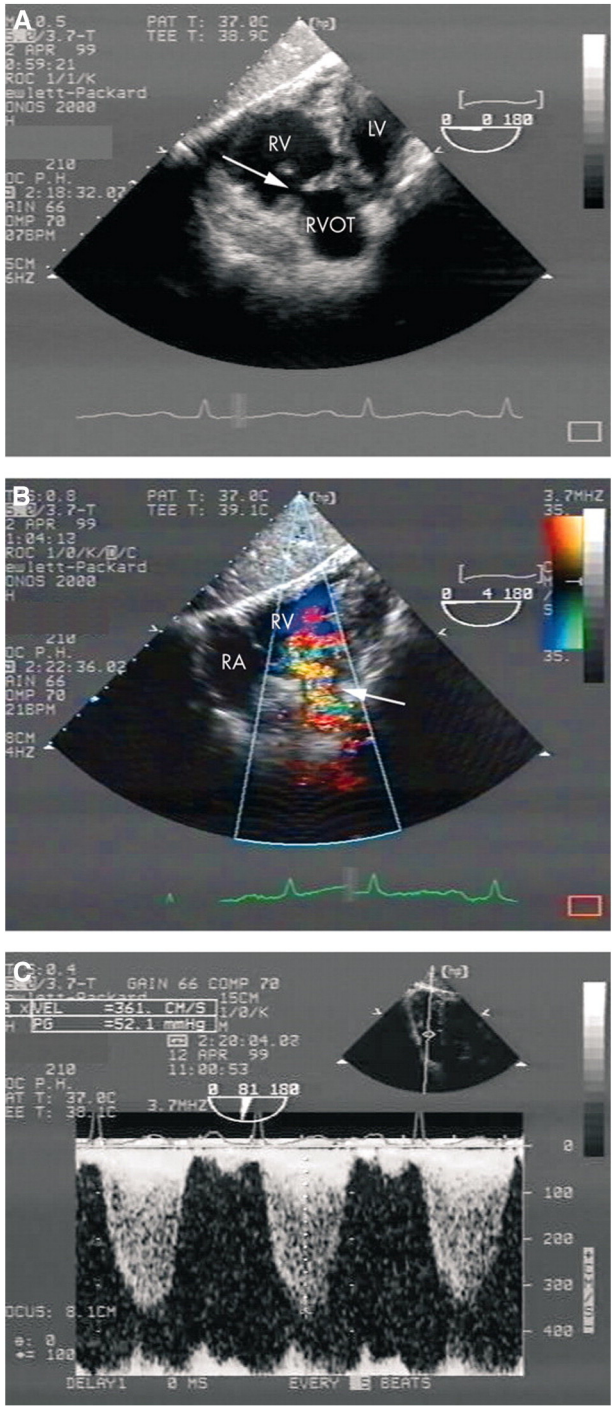

Fig. 4. (A) DCRV with low and discrete obstruction (arrow). Transesophageal echocardiography from within the fundus of the stomach in the horizontal plane (0°). (B) Same study plane, frontal view of the entire RV, end systolic frame. Color-coded flow Doppler imaging indicates origin of the turbulent flow within the right ventricular cavity (arrow); (C) Color-coded Doppler echocardiography allows proper quantitative evaluation of the mid cavitary pressure gradient of 52.1 mmHg by continuous wave Doppler. This also required rotation of the studying plane (81°). RA, right atrium. Reproduced from Hoffman et al. [2] with permission from *Heart*. Copyright 2004.

subcostal (transverse and frontal) and parasternal short axis planes have the most clinical value. However, since these views are especially difficult to achieve in adults, the use of transesophageal diagnostics is further emphasized [33].

Cardiac catheterization and angiocardiography can be used for confirmation of a diagnosis [23,24] (Fig. 5). In Hoffman et al.'s [2] study, cardiac catheterization was used to diagnose suspected DCRV when transesophageal ultrasound probes were not available; catheterization produced a successful diagnosis in all three cases. Bashore [24] suggested that cardiac catheterization may also be indicated in patients with anginal symptoms to rule out coronary artery disease. If correct placement is assured, catheterization is also a more accurate determinant of intraventricular pressure. Park et al. [11] warned that continuous Doppler may overestimate the true pressure gradient since it uses a simplified form of the Bernoulli equation that disregards velocity in the proximal ventricle.

Bashore [24] stated that transesophageal echocardiography is not necessary for diagnosis of DCRV but, instead, magnetic resonance imaging (MRI) is the best imaging modality (Fig. 6). Some studies have found that Doppler-echocardiography may be limited by poor imaging quality [24,36]. Singh et al. [37] acknowledged that while echocardiography is typically diagnostic, it may fail to show an anomalous muscle band, and MRI may be required to show a more exact anatomy in order to diagnose the pathology.

Park et al. [11] also used cinefluoroscopy and two-dimensional cine-magnetic resonance to identify DCRV. However, fluoroscopy is relatively limited in its ability to visualize structures, and neither study could determine intraventricular pressure gradients [11]. Compared to transesophageal echocardiography, neither of these methods alone can effectively distinguish true DCRV from other forms of outflow tract obstruction.

It is important to note that tetralogy of Fallot and DCRV with an associated ventricular septal defect may look similar on echocardiography. However, these can be easily distinguished since tetralogy of Fallot is characterized by decreased pulmonary flow whereas DCRV is associated with increased pulmonary flow [4].

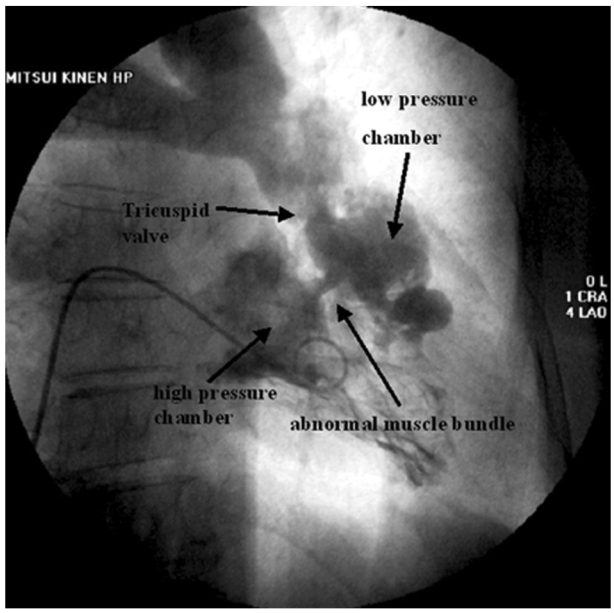

Fig. 5. Cardiac catheterization with angiography of DCRV. An abnormal muscle bundle separates the high-pressure inflow chamber and the low-pressure outflow chamber. Reproduced from Sawada et al. [34] with permission from *The Journal of Thoracic and the Cardiovascular Surgery*. Copyright 2005.

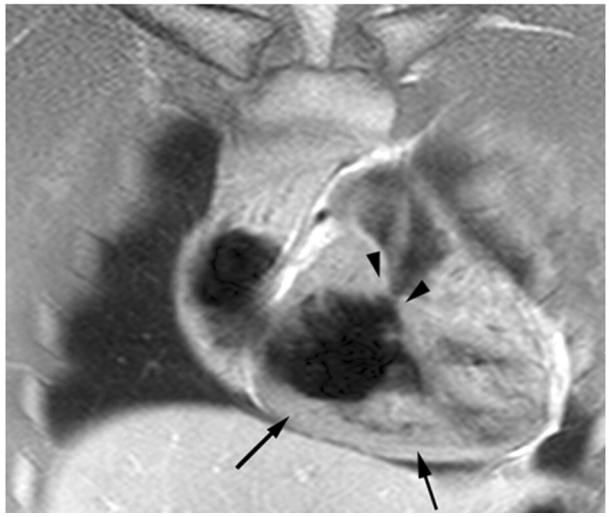

Fig. 6. Coronal T1-weighted MRI of the heart (repetition time msec/echo time msec, 2000/8.6; flip angle, 90°) shows hypertrophy of the wall of the RV (arrows) and an anomalous muscle band (arrowheads) dividing the RV. Reproduced from Kardux et al. [35] with permission from *Radiology*. Copyright 2007.

6. Treatment of DCRV

Once diagnosed, the most effective treatment of DCRV is surgical removal of hypertrophic or anomalous tissue using a right atriotomy [8,10,23,38]. However, Alva et al. [5] and Nagashima et al. [1] described a DCRV repair using a right ventriculotomy. Indications for surgery may include a pressure gradient over 40 mmHg between the proximal chamber and the pulmonary artery, aortic regurgitation, and symptoms of heart failure [1].

Given the anatomical location of the crista supraventricularis, care must be taken to preserve the normal attachments of the papillary muscles, as resecting too much tissue may result in postoperative dysfunction of the tricuspid valve [5,23]. Cil et al. [23] also warned of the difficulty in appreciating muscular anomalies intraoperatively if they are located low (toward the apex) in the RV. Such a position may make it difficult to appreciate the anomalous muscle as an outflow obstruction, especially if using a pulmonary trunk incision [23].

Conduction block is a potentially significant source of morbidity in the surgical treatment of DCRV. Alva et al. [5] reported that 12 of their patients (46%) were noted to have incomplete right bundle branch block postoperatively. This stands to reason since the right atrioventricular bundle runs along the septomarginal trabecula and moderator band as it courses towards the apex of the ventricle and to the anterior papillary muscle [15,16]. A partial or complete resection of the moderator band, therefore, may result in iatrogenic heart block [39].

Lucas et al., in 1962, described two cases of unrecognized DCRV, in which repair of what was thought to be a ventricular septal defect inadvertently closed the transventricular channel, with fatal results [31]. Such a report emphasizes the importance of correctly distinguishing outflow tract obstruction from other congenital anomalies. This is especially true in the case of ventricular septal defect, which is likely to be present with a DCRV. However, diagnosis of coexistent cardiac anomalies can be difficult, as they may be obscured by the origin of anomalous muscle bundles, as is commonly the case with ventricular septal defects [31].

Since isolated DCRV is rare, resection of obstructive tissue is often accomplished concomitantly with repair of associated congenital disease. Loukas et al. [16] reported that the location of the moderator band in the morphological RV poses difficulties during repair of apical ventricular septal defects. This has particular significance in the

setting of DCRV since hypertrophy of the crista supraventricularis might further reduce access to the ventricular septum.

In the event that a patient does present without significant coexisting defects, surgical intervention might not be necessary until a pressure gradient of 40 mmHg is observed [2]. Tamai et al. [40] reported a case of a 71-year-old woman diagnosed with DCRV complicated by severe calcification of the tricuspid valve. The calcification was believed to result from high pressure outflow in the RV, which was supported by findings of a high velocity flow signal on Doppler echocardiography [40]. If a patient has mild symptoms and refuses surgery, beta-blocker therapy may be sufficient to improve their general condition [29,38]. Tamai et al.'s [40] patient was successfully treated with carvedilol after refusing surgical treatment.

Since DCRV is generally a progressive pathology, it may still be beneficial to operate before the emergence of symptoms [33]. Disease groups at high risk for DCRV include those with ventricular septal defects, increased pulmonary flow, and rapid-onset or primary right heart failure without associated pulmonary hypertension [4,10,11]. Kardux et al. [35] reported a case in which a 17-year-old male presented with dyspnea and a history of pulmonary valve endocarditis. He was diagnosed with DCRV using electrocardiogram and MRI, and his history of endocarditis was attributed to increased pressure flow over the right ventricular outlet tract causing damage to the pulmonary valve. Lopez-Pardo et al. [28] had earlier described a similar case of DCRV complicated by mural and pulmonic valve endocarditis. This was further complicated by a septic pulmonary embolism requiring surgical intervention. Cases such as these underscore the importance of surgical correction of DCRV, particularly in young patients, before serious complications may arise. Wilder et al. [41] recently reported a case of a 31-year-old woman diagnosed with DCRV whose only presenting symptom was a nonproductive cough. She was found to have pulmonic valve and RVOT endocarditis with a pulmonary nodule secondary to embolization. This case also demonstrated the importance of surgical intervention, even when patients are not particularly symptomatic. Telagh et al. [38] reported excellent results following surgical repair of DCRV in 35 patients, both pediatric and adult, spanning a period of 18 years. They performed resections of anomalous muscular bundles, enlarging the RVOT, and reported no related deaths, no postoperative arrhythmias, and no need for long-term medical therapy. Kveselis et al. [42] reported similar findings. They found that mild sequelae such as low-grade murmur or mild tricuspid regurgitation were common, but serious cardiac complications were very rare following surgical repair of DCRV.

Pulmonary balloon valvuloplasty has been used as an alternative to tissue resection for pulmonary valve stenosis. Cil et al. [23] reported a decrease in the ventricular pressure gradient by 35–50% in four patients. In two of those patients, the treatment was ineffective in abolishing the gradient entirely and required follow-up surgery.

7. Conclusions

DCRV is a developmental cardiac defect, in which the RV is divided into a high-pressure proximal and low-pressure distal chamber. It can be caused by anomalous muscle tissue, the hypertrophy of endogenous trabecular bands, or by an aberrant moderator band. DCRV is typically found concomitantly with congenital cardiac disorders, most notably ventricular septal defect and subaortic stenosis [2]. Due to its rarity and the difficulty of visualization, DCRV continues to be misdiagnosed [2,23,28] . The most effective diagnostic studies are transesophageal echocardiogram [2], continuous flow Doppler, and cardiac catheterization. Surgical excision of obstructive tissue is the most effective form of treatment.

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